CASE REPORTS

REVIEW OF PRIMARY TUMORS OF THE DIAPHRAGM

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A case report of a man with a huge, left-sided diaphragmatic rhabdomyosarcoma is presented. The mass was attached to the lung, pericardium, left phrenic nerve, sternum, and chest wall. En-bloc resection of the tumor was done with primary repair of the diaphragmatic defect. Only three reported cases of primary diaphragmatic rhabdomyosarcoma were found in the literature.

Primary tumors of the diaphragm are quite rare. Until 1968, only 85 cases had been reported in the world medical literature.[1] Of those 85 cases, 52 were benign tumors and 33 were malignant (Table 1). Cystic lesions and lipomas were the two most common benign lesions, and fibrosarcoma was the most frequent malignant lesion. Wiener and Chou[2] reported two cases of rhabdomyosarcoma of the diaphragm in 1965. Since that time, no other cases have been reported in the English language. Recently, a patient suffering from primary diaphragmatic rhabdomyosarcoma was treated. A description of this case, as well as a collective review of the English literature on primary diaphragmatic tumors, is presented in this report (Table 1).[3-11]

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CASE REPORT

A 58-year-old white man was admitted to the King/Drew Medical Center on February 11, 1980, with abnormal chest film findings. The patient complained of anorexia and weight loss of 30 lb in the prior two months without pulmonary symptoms. Medical history was not significant. Physical examination revealed dullness over the left lower chest, with decreased breath sounds over the same area.

Laboratory examination showed a red blood cell count of 2.85 mil/mm^3; hemoglobin 7.5 mg/dL; hematocrit 23.5 percent; and increased liver enzymes and prothrombin time. Arterial blood gases on room air revealed pO_2 81 torr; pCO_2 38 torr; pH 2.44; and oxygen saturation 96 percent. Pulmonary function tests indicated moderate restrictive lung disease. Chest film showed a large, round, homogeneous density in the left lower lung field. Abdominal ultrasonogram indicated an 11 × 11 cm mass in the region of the left hemidiaphragm. Intravenous pyelogram, liver-spleen scan, and barium enema disclosed no abnormalities in these organs. A Gallium[6,7] scan revealed an increased uptake in the left upper quadrant of the abdomen.

Exploratory left posterolateral thoracotomy was performed through the 6th rib bed and a large ovid mass, originating from the left diaphragm, was removed. The tumor mass, well encapsulated by a pseudocapsule, was firm in consistency. The upper pole of the mass was attached to the inferior surface of the left lower lobe of the lung. Medially, the surface of the tumor was attached to the anterolateral surface of the pericardium and the left phrenic nerve; and, anteriorly, the surface of the tumor was attached to

TABLE 1. PRIMARY TUMORS OF THE DIAPHRAGM

Pathologic Diagnosis	Collected by Weiner and Chou[2] 1868–1963	Collected by Olaffson[1] 1963–1968	Collected by Authors 1969–1982*	Total
Benign				
Angiofibroma	3	1	0	4
Adenoma, adrenal cortical	1	0	0	1
Adenoma, liver cell	1	0	0	1
Chondroma	1	0	0	1
Cyst	12	6	1 (Greenberg)[3]	19
Fibroma	3	1	0	4
Fibrolymphangioma	1	0	0	1
Fibromyoma	1	0	0	1
Hamartoma	1	1	0	2
Lymphangioma	1	0	0	1
Leiomyoma	1	0	0	1
Lipoma	9	0	4 (Kalen)[4]	28**
Mesothelioma	1	0	0	1
Neurilemmoma	2	0	1 (Sarot)[5]	3
Neurofibroma	4	0	0	4
Rhabdomyofibroma	1	0	0	1
Malignant				
Chondrosarcoma	0	0	1 (Ujiki)[6]	1
Embryonal carcinoma	0	0	1 (Fortner)[7]	1
Fibroangioendothelioma	1	0	0	1
Fibromyosarcoma	2	0	0	2
Fibrosarcoma	8	1	2 (Sbokos)[8] (Walsh)[9]	11
Hemangioendothelioma	1	1	0	2
Hemangiopericytoma	1	0	1 (Seaton)[10]	2
Leiomyosarcoma	1	0	1 (Dionne)[11]	2
Myosarcoma, myoblastic	2	0	0	2
Neurofibrosarcoma	2	0	0	2
Rhabdomyosarcoma	2	0	1 (Authors)	3
Sarcoma	2	1	0	3
Sarcoma, endothelial, vascular	2	0	0	2
Sarcoma, mixed cell	2	0	0	2
Sarcoma, undifferentiated	1	1	0	2
Synoviosarcoma	1	0	0	1
Mesenchymoma	0	1	0	1
Total	71	14	13*	113**

* Only in English literature
** Included Kalen's review of world literature

the sternum and chest wall, but there was no invasion of the bony thoracic cage. No gross lymph node involvement was noted. En-bloc removal of the tumor mass, including a portion of the pericardium, the phrenic nerve, the anterior parietal pleura with the endothoracic fascia, and the diaphragm was accomplished. The diaphragm was excised with a free margin approximately 2 cm from the tumor. The diaphragmatic defect was repaired primarily in two layers by using a continuous 0-Dexon suture. Three interrupted horizontal matress sutures, over a Teflon pledget, using 00-Tevdek, were applied over the central portion of the suture line.

The resected specimen consisted of a large, ovid, solid, lobulated tumor measuring 15 × 10 × 12 cm, and weighing 800 g. The tumor was partly covered by a tan-gray fibrous capsule. Along the middle of the outer surface ran an elevated ridge halfway across the mass, comprising the diaphragmatic attachment. The cut surface showed a soft, firm, white-gray to

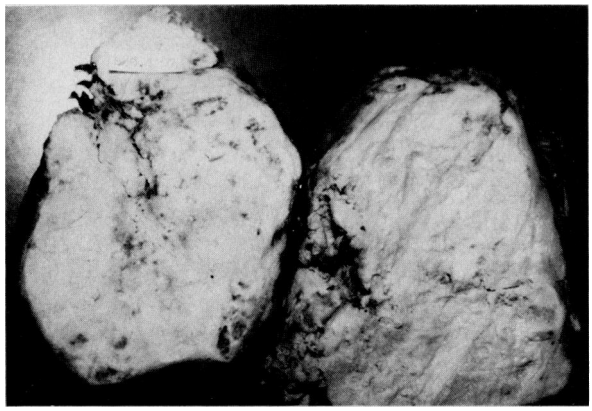

Figure 1. Bisected diaphragmatic tumor showing solid areas with necrosis and cyst formation

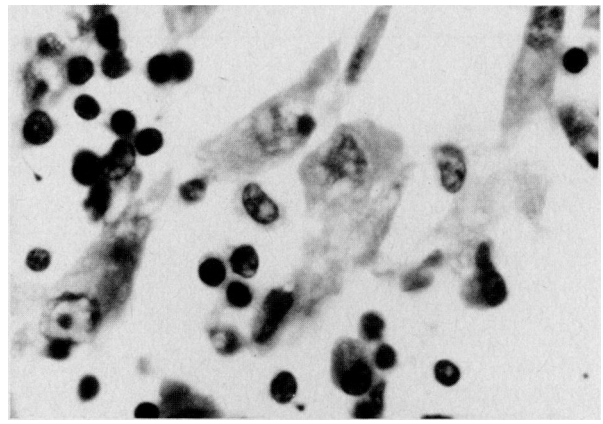

Figure 2. Photomicrograph of the rhabdomyosarcoma showing pleomorphic cells with abundant cytoplasm and large vesicular nuclei (H & E, ×1000)

pink-tan solid tumor with areas of necrosis and pseudocystic formation (Figure 1).

Microscopically, the tumor consisted of solid sheets of pleomorphic neoplastic cells containing abundant eosinophilic cytoplasm, and large vesicular nuclei with prominent nucleoli, hyperchromasia, and mitoses (Figure 2). Some of the large cells were multinucleated (Figure 3). The tumor was classified as pleomorphic rhabdomyosarcoma.

After surgery the patient did well and was extubated on the first postoperative day. On the sixth postoperative day, however, he developed adult respiratory distress syndrome. He died from respiratory failure 37 days postoperatively.

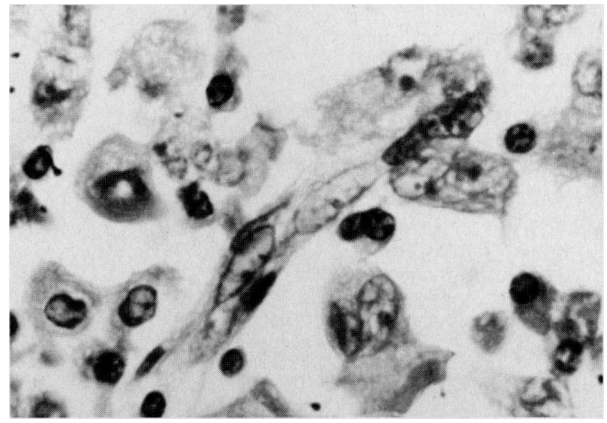

Figure 3. Photomicrograph of another field of the neoplasm showing multinucleated cells (H & E, ×1000)

DISCUSSION

Primary diaphragmatic tumors occur with approximately the same rate in both sexes. In about three quarters of all patients, the tumor is detected on routine chest x-ray films, without specific symptoms or radiologic characteristics.

Radiographic evaluation includes chest film, upper gastrointestinal series, barium enema, intravenous pyelography, angiography, liver-spleen scanning, bronchography, and pneumoperitoneum to delineate and distinguish the mass from other lesions.[12] Positive-contrast peritoneography was introduced by White and coauthors[13] to detect diaphragmatic abnormalities in infants, but was not reported as a diagnostic tool of diaphragmatic tumors. Abdominal ultrasonography is useful to differentiate between a solid mass and a cystic mass; and computed axial tomography will probably prove to be one of the best radiologic examinations to delineate the mass and rule out other pathology or extension of the tumor into other organs. Thoracoscopy and biopsy can establish the diagnosis, especially for suspected lipomas; but most authors prefer thoracotomy to establish and correlate the diagnosis of diaphragmatic tumors with the treatment.

Various methods of closing the defect have been reported, chiefly in animal experiments. Transplantation of the diaphragm, the chest wall flaps, and the fascia lata with human dura and various prosthetic materials have been used both experimentally and clinically. The procedure of choice for closing a large diaphragmatic defect depends upon individual preference, but it will be better to use prosthetic material than an autogenous tissue. Prosthetic material is more

readily available and requires a less extensive procedure. Touloukian[14] reported the incorporation of skeletal muscle into the silastic sheet used for diaphragmatic repair on pups; but Lacey and coauthors[15] published a case of unsatisfactory repair of an agenesis of the diaphragm using the silastic sheet without evidence of skeletal muscle ingrowth into the material. Failure of skeletal muscle ingrowth in their case may stem from the absence of diaphragmatic muscular incorporation into the silastic sheet. Polypropylene mesh, which is firmly incorporated by fibroblastic ingrowth, will be the better material until it is clinically or experimentally proven that the silastic sheet is incorporated by muscular ingrowth in adults.

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